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*Case Report*

# Intracranial haemangioma: clinical features and radiological appearances

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Since the advent of magnetic resonance imaging (MRI), intracranial cavernous haemangioma has become the most commonly identified occult vascular malformation. The following case report demonstrates the characteristic radiological appearances and highlights the central role of MRI in the diagnosis of this lesion.

*CASE REPORT* A thirteen-year-old boy presented with a six-week history of a progressive left hemiparesis, with no associated symptoms. Clinical examination revealed an upper motor neurone weakness of the left arm and leg. Examination was otherwise unremarkable. Computed Tomography (CT) (Figure 1) and MRI (Figure 2) of brain were performed.

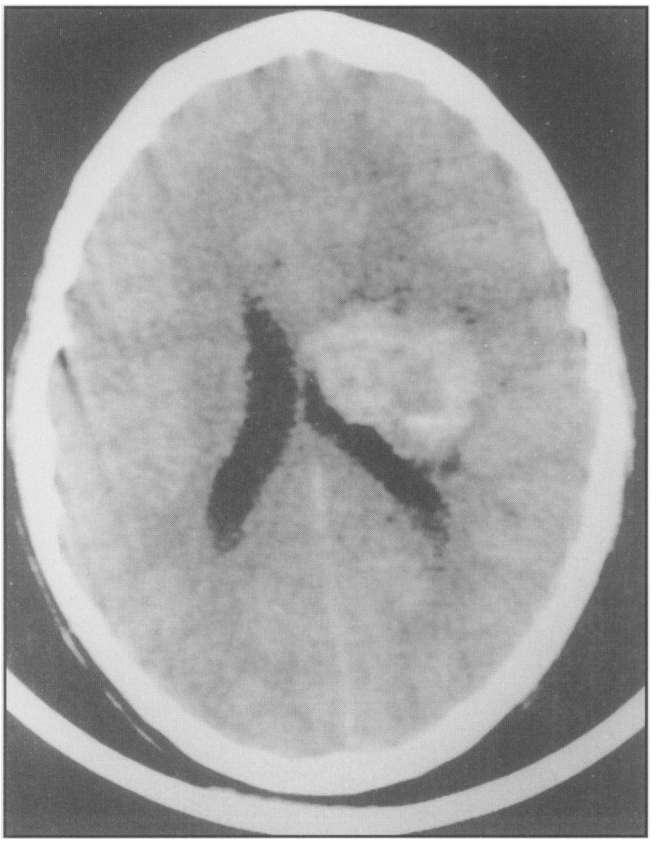

*Fig 1.* Axial CT scan (unenhanced).

The CT images demonstrate an irregular, mainly hyperdense lesion in the periventricular white matter adjacent to and causing compression of the body of the right lateral ventricle. Foci of high attenuation, consistent with calcification, are present within the lesion, and a surrounding low density zone of oedema is noted.

The lesion is seen on MRI to extend superiorly into the centrum semiovale and inferiorly into the upper basal ganglia on the right. It demonstrates a 'popcorn' appearance, with a reticulated core of mixed signal intensities. Appearances are typical of a cavernous haemangioma containing haemorrhage in various stages of evolution.[1] A peripheral rim of low signal on the T2 weighted

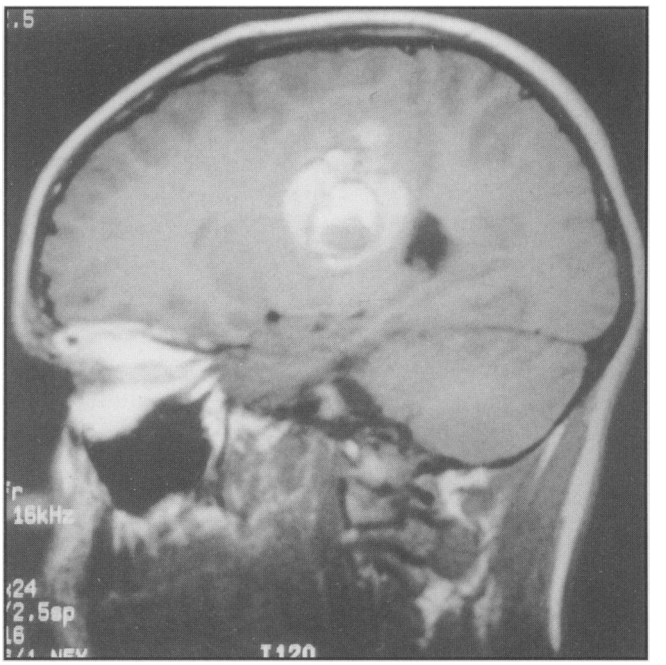

*Fig 2a.* Sagittal T1 weighted (TR500/TE11) image

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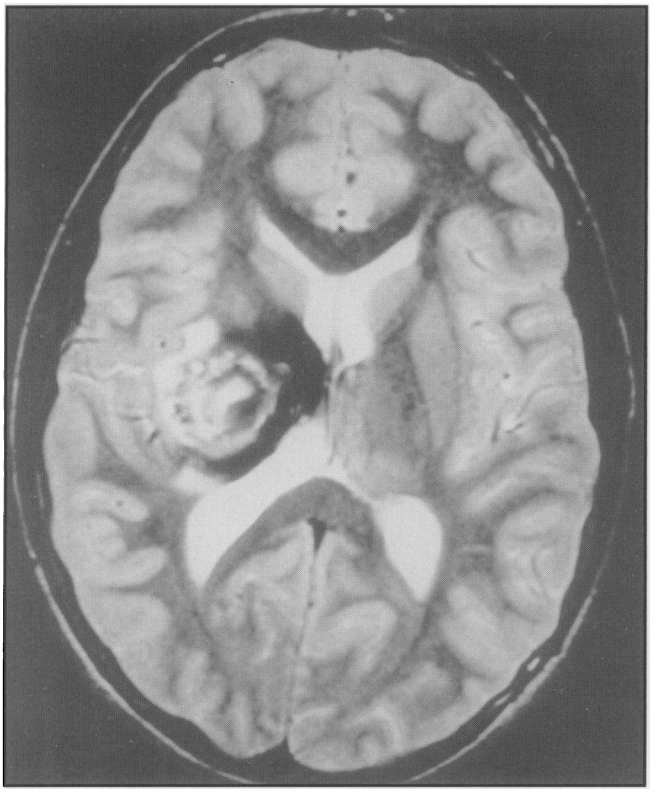

*Fig 2b.* axial T2 weighted (TR4000/TE84) image

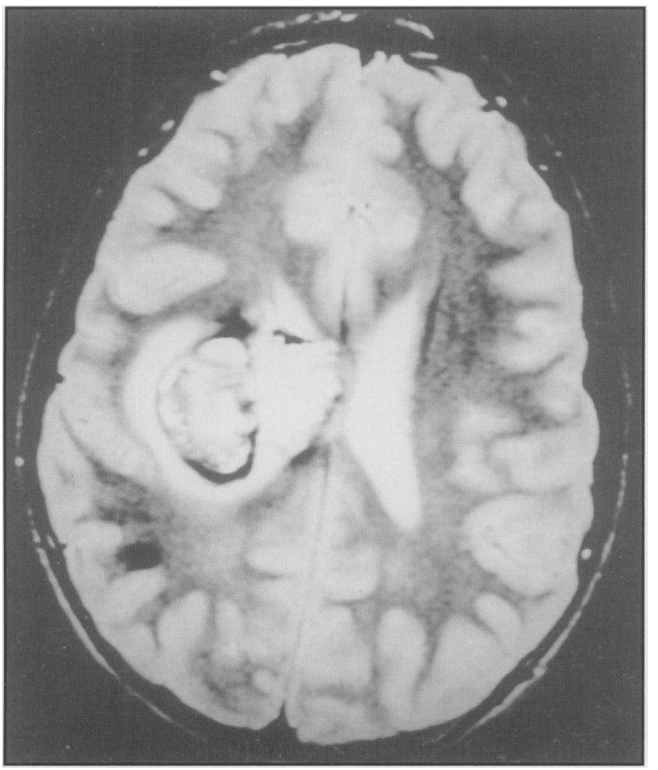

*Fig 2c.* Axial T2 weighted image

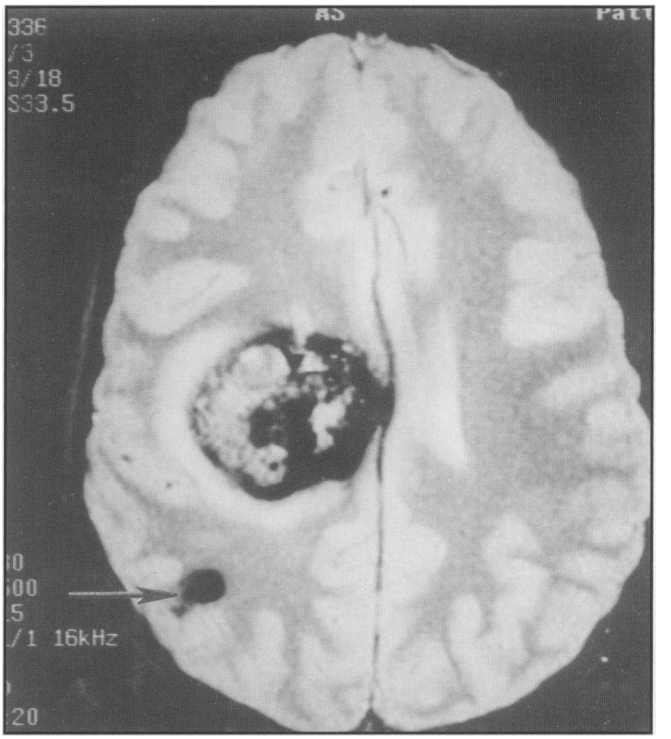


*Fig 3a.* Gradient echo acquisition (TR500/TE15 flip angle 30°) images. The lesion is of lower signal than on the T2 weighted images and the presence of a second smaller lesion (arrow) which is just visible on the T2 weighted images (figure 2c) is confirmed.

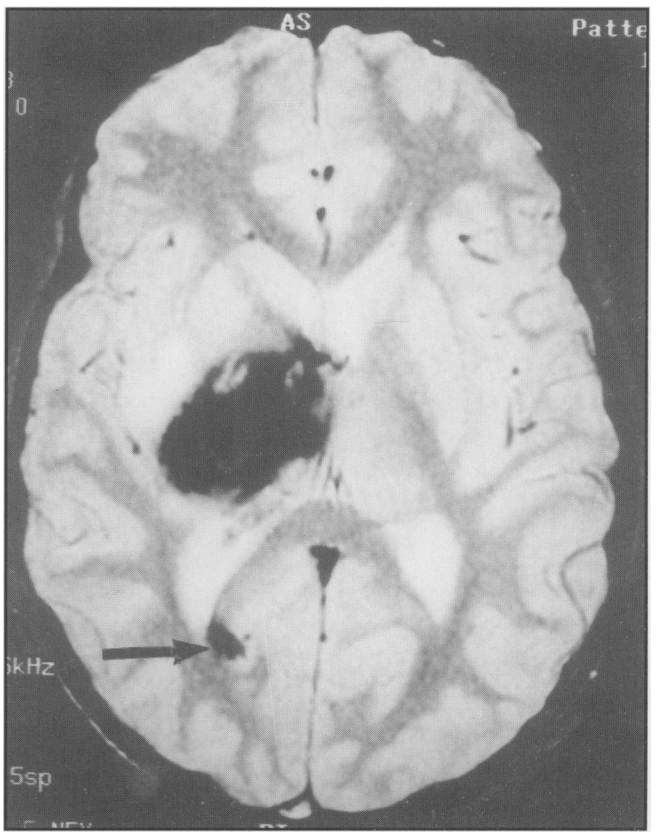


*Fig 3b.* a third lesion (arrow), not visible on T2 images, is identified.

images represents haemosiderin. A second lesion, not visible on CT, is seen in the subcortical white matter of the right parietal lobe (Figure 2c).

On subsequent gradient echo (GRE) acquisition images the lesion appears more hypointense and the peripheral haemosiderin rim is much more pronounced (Figure 3a). The second lesion is more apparent on the GRE images, which also reveal a third lesion adjacent to the right occipital horn. The diagnosis is supported by the presence of multiple lesions, seen in 50% of all cases.[1]

The largest lesion was surgically evacuated with subsequent clinical improvement. The diagnosis of cavernous haemangioma was confirmed on histology.

**DISCUSSION**

Vascular malformations of the central nervous system are congenital lesions which are classified into five anatomic types, according to the abnormal vessels identified on histology.[2] These include (1) telangectasias, (2) varices, (3) cavernous malformations, (4) arteriovenous malformations, (5) venous malformations. Cavernous haemangiomas account for 5-13% of all vascular malformations[3] and, with the advent of MRI, are now the most commonly identified angiographically occult vascular malformation,[4] with a reported incidence of 0.47%.[5]

There is a familial form of the disorder, which may be inherited as an autosomal dominant trait with variable penetrance, and the causative gene lies on the long arm of chromosome 7.[6, 7] The familial form is characterized by multiple lesions, occuring in 50-73% of cases, while multiplicity of lesions is found in less than 33% of sporadic cases.[3]

The MR pattern reflects the histological composition of the cavernous haemangioma, which is of a closely approximated collection of endothelial-lined vascular spaces whose walls are composed of collagen and devoid of smooth muscle and elastica, with virtually no intervening normal neural tissue. Haemorrhagic material of varying age is present within the lesion, and there is haemosiderin staining of the surrounding brain.[3]

78% of these angiographically occult lesions are supratentorial,[5] occurring most commonly in the frontal and temporal lobes. Common posterior fossa sites include the pons, cerebellum and medulla. Lesions are also described in the spinal cord and in extra-axial sites.

When symptomatic, presentation most commonly occurs between 20 and 40 years of age with seizures, focal neurological deficit or headaches.[5] Male patients are more likely to present at a younger age (<30years), and often with epilepsy, while female patients usually present at 30-60 years and are likely to have gross haemorrhage and greater neurological deficit.[3] Paediatric patients also have a greater likelihood of overt haemorrhage. Familial lesions tend to be more aggressive and successive generations manifest symptoms at earlier ages.[3]

Frequent occult haemorrhagic episodes occur with a 0.7% annual risk of an overt bleed.[5] The initial bleed is usually self-limiting but there is increased risk of recurrent haemorrhage (14-29%),[8] which is often more severe. This is associated with progressive neurological decline and severe residual deficit. This is particularly significant in the brainstem, where the typical pattern of exacerbation and remission of symptoms results in much greater neurological deficit than normally occurs in supratentorial lesions.

Management strategies must balance the risk of treatment against the natural risk. This is related to the clinical presentation and the site of the lesion. Asymptomatic patients require only clinical follow-up and MRI. The current established indications for surgery are overt haemorrhage, focal neurological deficit and/or intractable epilepsy.[3] This is straightforward for symptomatic lesions in accessible sites. In the case of a more deepseated lesion, e.g. basal ganglia or thalamus, the risks must be very carefully assessed as the risk of surgery is obviously much greater. Radiosurgery may be of value in this situation.[3] Overall results following surgery are generally favourable. A review of many series estimating clinical outcome reports a fair to excellent outcome in 76-100% of patients, with a poor outcome in <19%.[3] Good results have also been reported for surgery of brainstem[9, 10] and spinal cord lesions.[11]

Intracranial haemorrhage demonstrates specific spin echo (SE) intensity patterns, based on the evolutionary stage of the haemorrhage and therefore on the paramagnetic blood breakdown products present.[1, 12] Intracellular deoxyhaemoglobin, present in the acute phase, does not affect T1 relaxation times and therefore appears isointense on T1 weighted images. Intracellular

methaemoglobin appears in the early subacute phase and causes T1 shortening and resultant high signal.

The above substances produce static field inhomogeneities due to their magnetic susceptibility effects, resulting in shortening of T2 relaxation times. They therefore appear hypointense on T2 weighted images. However in the late subacute phase when methaemoglobin is released from disrupted red blood cells, increased signal is seen, possibly due to associated fluid accumulation. In the chronic phase, intralysosomal haemosiderin appears at the edges of the haematoma. This does not affect T1 but produces T2 shortening and reduced signal on T2 weighted images.

GRE studies are more sensitive than routine SE techniques to acute and chronic haemorrhage as T2* effects are present (there is no 180° refocussing pulse). Shortening of T2* relaxation times produces more marked hypointensity in areas of haemorrhage and a more prominent haemosiderin rim. The specificity of MR findings is less with GRE than with conventional SE images, because of the marked hypointensity,[13] as specific lesion signal characteristics may be lost. Neither is the hypointensity specific for haemorrhage (also caused by calcium, ferritin, melanin and air). The best use of GRE studies is therefore as an adjunct to SE sequences.

MRI is therefore the imaging modality of choice in the diagnosis of intracranial cavernous haemangioma. Both GRE and routine SE sequences should be performed to optimise visualisation of intracranial haemorrhage and detect multiple lesions and therefore maximise the sensitivity of the investigation.

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